

Case report

Diabetic amyotrophy in a teenage boy

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Diabetic amyotrophy is a rare complication of diabetes mellitus at any age, but it is almost unreported in the literature below the age of 30 years. We report the case of a 16 year old boy who attempted suicide due to the symptoms of diabetic amyotrophy.

CASE REPORT. A 16-year-old boy was admitted following an overdose of 48 tablets of diclofenac 100 mg, 20 tablets of codeine phosphate 30 mg and 40 tablets of paracetamol 500 mg. He was alert and felt well on admission. General examination was normal. His serum paracetamol level on arrival was 34 mg/l, which did not justify treatment with acetyl cysteine. Serum sodium, potassium and urea were normal. He was treated conservatively and observed overnight. On further questioning the patient revealed that the reason for his overdose was increasing clumsiness in his legs over the preceding six months which had halted a promising athletic career and had led to him being the focus of teasing at school. His home situation seemed to be supportive and caring, and no other cause for his attempted suicide was apparent.

He admitted to polyuria and polydipsia associated with weight loss of approximately 5 kg over the six months prior to admission. Detailed examination of his central nervous system revealed normal cranial nerve function, with normal optic fundi. A grade 2/5 weakness in his right extensor hallucis group of muscles was present, but no obvious weakness of the quadriceps or psoas muscle groups was detected, despite wasting of both thighs and calves. His gait was clumsy and broad-based, with hyper-extension of both knees. He was unable to stand on tiptoes. Tendon reflexes were absent in arms and legs, with downgoing plantar responses. There was no sensory deficit or impairment of joint position sense.

His random plasma glucose was 33.1 mmol/l. His haemoglobin concentration, white cell count, serum urea electrolytes and liver function tests were normal. Serum thyroxine, thyroid stimulating hormone, vitamin B12 and folate, complement, immunoglobulins and immune complexes were all normal. Serum auto-antibody tests, rheumatoid factor and porphyrin levels were normal. His serum aldolase was 3.5 iu/l (normal range 1.2 to 7.6) and creatine kinase 40 iu/l (less than 150). Radiograph of the lumbar spine was normal.

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The diagnosis of diabetes mellitus was confirmed by the hyperglycaemia, and treatment was started with insulin. Within two weeks his plasma glucose levels were well controlled and he was allowed home. He was reviewed two weeks, six weeks, and four months later to assess the progress of both his diabetes and his muscle weakness. Exercise tolerance, assessed at two weeks and at four months by maximum running distance in ten minutes had increased from 760m to 1765m. By the end of four months the wasting of his legs noted on admission had resolved (thigh measurements increased from 41 to 49 cms and calf measurements from 15 to 25 cms), and his tendon reflexes had returned to normal. His diabetic control was good with average plasma glucose throughout the day 7.5 mmol/l and glycosylated haemoglobin (HbA1) 9.3% (normal <7.5%). He felt considerably better in himself and all depressive and suicidal ideas had gone.

DISCUSSION

Diabetic amyotrophy is an unusual presentation of diabetes mellitus, as too is attempted suicide. First described in 1890 by Bruns in Germany it was not until 1953 that Garland¹ again brought the condition to the notice of contemporary clinicians. Described as a disease associated with poor control of diabetes mellitus^{2, 3} it usually presents with weakness and muscle wasting of the lower limbs, often associated with pain, and usually without loss of touch or joint position sense.² It is more common in the older diabetic and we have been able to find only one report of a patient under the age of 33 years in the past seven years.³ The absence of pain in the legs does not fit with the more usual presentation of the condition but there are examples in the literature of pain-free diabetic amyotrophy.^{2, 4} The involvement of the extensor hallucis group of muscles is of interest as Garland described a similar distribution in three of his 12 cases.² The term diabetic proximal motor neuropathy is sometimes preferred but is less appropriate in this case as there was no demonstrable proximal weakness. The abnormal gait and hyper-extension of the knees would, however, indicate some involvement of the quadriceps and psoas muscle groups.

The pathogenesis of diabetic amyotrophy remains obscure. Garland felt on the basis of nerve conduction studies that the primary lesion was in the lumbar spinal cord and he postulated a lower motor neurone neuropathy.¹ It was noted that as the diabetic control improved so also did the muscle weakness.^{2, 3, 4} This is consistent with the metabolic theory, with accumulation of intraneuronal sorbitol and depletion of myoinositol. This results in nerve dysfunction and damage.^{5, 6, 7} These metabolic pathways are dependent on the enzyme aldose reductase, and the dysfunction and biochemical abnormalities of diabetic neuropathy can be improved by the administration of aldose reductase inhibitors.^{5, 6} Diabetic neuropathy is usually diffuse and symmetrical but there are often focal features, particularly in diabetic amyotrophy. This suggests that microvascular disease contributes to pathogenesis. The demonstration of resistance in a diabetic nerve to the effects of ischaemia, possibly due to an adaptive process, is further evidence of the vascular theory.⁷ The precise aetiology, however, remains unclear, but a combination of factors is likely.

The differential diagnosis would include femoral neuritis, sciatica, carcinomatous neuropathy, intra-pelvic neoplasm, motor neurone disease, thyrotoxicosis and

Guillain-Barré syndrome. Electrophysiological studies and a lumbar puncture would have helped to confirm the diagnosis, but we are satisfied that our investigations, and the marked improvement with control of his diabetes, indicate that the correct diagnosis of this case is diabetic amyotrophy.

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